TITLE:

脊椎・脊髄領域における造影剤投与による3次元核磁気共鳴画像法の有用性に関する研究(Dissertation_全文)

AUTHOR(S):

Okumura, Ryosuke

CITATION:

Okumura, Ryosuke. 脊椎・脊髄領域における造影剤投与による3次元核磁気共鳴画像法の有用性に関する研究. 京都大学, 1994, 博士(医学)

ISSUE DATE:

1994-01-24

URL:

https://doi.org/10.11501/3093807

RIGHT:

主論文1は出版社の許諾条件により公開していません

京都大学学術情報リポジトリ
KURENAI 紅
Kyoto University Research Information Repository

脊椎・脊髄領域における造影剤投与による3次元核磁気共鳴
画像法の有用性に関する研究

奥村亮介

脊椎・脊髄領域における造影剤投与による3次元核磁気共鳴
画像法の有用性に関する研究
奥村亮介

CF-A4SN 70

②

主論文 1

Three-dimensional MR Vascular Imaging of the Spine Using Gadolinium-DTPA

Ryosuke Okumura, M.D., Reinin Asato, Doc. Med. Sci., Yukio Miki, M.D., and Junji Konishi, Doc. Med. Sci.

Reprint from Radiation Medicine Vol. 10 No. 6
November–December 1992

主論文 2

Epidural Venous System (Meningorachidian Venous Plexus) in Juvenile Amyotrophy of Distal Upper Extremity; Assessment with Gadolinium-DTPA Enhanced Volumetric MR Study

Ryosuke Okumura[1], Reinin Asato[1], Hidenao Fukuyama[2], Masahumi Ogawa[2], Yukio Miki[1], and Junji Konishi[1]
Department of [1]Nuclear Medicine and [2]Neurology, Faculty of Medicine, Kyoto University.

Address correspondence and reprint requests to:
Ryosuke Okumura at the Department of Nuclear Medicine, Faculty of Medicine, Kyoto University, 54 Shogoin Kawahara-cho, Sakyo-ku, Kyoto 606-01 Japan.
Phone: 81-75-751-3742
FAX: 81-75-771-9709, 81-75-752-3144

Abstract

The epidural venous system (meningorachidian venous plexus) was analyzed utilizing Gadolinium-DTPA (Gd-DTPA) enhanced volumetric MR images in eleven patients with focal cervical spinal cord atrophy, clinically consistent with juvenile amyotrophy of distal upper extremity.

In our series, all of the patients showed unusual posterior epidural venous enhancement at the C5-6 level, suggesting posterior epidural venous dilatation. Three patients also showed prominent dilatation of cervico-thoracic epidural veins surrounding the thecal sac. These MR findings were also demonstrated by spinal phlebography. Gd-DTPA enhanced MR images, especially high resolutional volumetric MR images, were efficient for evaluating these vessels. The observation of meningorachidian venous plexus along the disease course should be necessary for searching the pathogenesis of this disease.

Key Words; Spinal Cord - diseases, Veins, Magnetic Resonance Imaging NMR

Introduction

Juvenile amyotrophy of distal upper extremity is a clinical disease entity first published by Hirayama et al. in 1959 (1). This disorder has unique clinical features; juvenile onset, male preponderance, and characteristic distal and segmental muscular atrophy in the hand and forearm, most often unilateral. The muscular atrophy rapidly progresses during the first 2 to 3 years after insidious onset, and thereafter shows a stable state. There is no sensory disturbance or involvement of cranial nerves and pyramidal tracts. Hence, this benign prognosis differs entirely from that of motor neuron disease (2-9). However, the pathogenesis of this muscular atrophy is still unknown. Histopathological study of a patient with this disorder shows selective loss of spinal anterior horn motor neuron, most prominent in the lower cervical region (10). Spinal cord atrophy is noted within several years after onset (11).

Radiological findings concerned with epidural veins were reported in reference to this disorder. Iwasaki et al. stated that the posterior part of the dural canal becomes tense and shifts forward in the lower cervical region due to pressure on the dural canal by the bony spinal canal in flexed neck position (12). Recurrent pressure accompanied with neck movement provokes posterior epidural space widening and abnormal posterior epidural venous dilation. This hypothesis, called flexion myelopathy, suggests that the dilatation of the posterior epidural vessels plays an important role in spinal cord atrophy (12,13). On the other hand, Aii et al. reported Gadolinium-DTPA (Gd-DTPA) enhanced MR images showing prominent epidural enhancement surrounding the thecal sac, suggesting dilatation of whole cervical epidural veins in this disorder. The dilated cervical epidural veins were operatively confirmed, and the histopathological findings of epidural tissue strongly suggested venous malformation (14).

In the present study, the authors analyzed the epidural veins in juvenile amyotrophy of distal upper extremity with the Gd-DTPA enhanced volumetric MR imaging (15) and spinal phlebography (16,17).

Materials and Methods

Eleven patients (8 males and 3 females, ages ranging from 16 to 48 years, with average of 23 years), with clinical history and physical findings compatible with juvenile amyotrophy of distal upper extremity, were studied. Deep tendon reflexes were depressed on the affected extremities and no sensory involvements were detected clinically. Atrophy of the muscle was distributed in the spinal segments. All patients showed anterior horn cell damage on EMG, corresponding to localized muscular atrophy. Nerve conduction studies disclosed no abnormalities, including F-wave. All patients except one had less than 4 years of history of motor weakness of unilateral upper extremity, and were considered to be in an evolutional or early stabilizing phase of the disease (11). One patient (case 8) was studied 30 years after the onset (Table 1).

The examinations were performed on a 1.5 Tesla MR unit (Signa, GE, Milwaukee, WI, U.S.A.), using a posterior cervical surface coil. The T1-weighted images were obtained in the sagittal and axial planes, with a repetition time (TR) of 600 msec and an echo time (TE) of 20 msec. The volumetric scan was performed immediately after the intravenous injection of gadolinium diethylenetriamine penta-acetic acid (Gd -DTPA) (0.1-0.15 mmol/kg BW). The scan method employed was a time-of-flight technique (sagittal volume fast scan, 1.9 mm partitional thickness, 28 slices, TR/TE = 100 msec / 16 msec, flip angle of 45 degrees, 256 x 256 matrix, one averaging), with first-order gradient moment nulling. Scanning time was approximately 14 min (15).

The patients' neck position was neutral to slightly flexed along the curvature of the posterior cervical coil. In five cases (cases 6,7,and 9-11), T1-weighted images were also obtained in neck flexed position.

The epidural veins were evaluated in sagittal partitional, or axial and coronal reformatted images. Phlebographic projection images were obtained by collapsing several partitional images, using the maximum intensity projection method. Sizes of posterior epidural veins were evaluated on reformatted axial images at the C5-6 level. The asymmetry of internal jugular veins was also evaluated on T1-weighted axial images at the C6 level. Conventional spinal phlebography was conducted in four cases (cases 1,2,4 and 6) and was used as reference information for the study of epidural veins.

Results (Table 2)

All patients showed focal spinal cord atrophy (C5-Th1 level), and 9 of them showed typical hemiatrophy at the C5 - 6 level.

Eight patients (cases 4-11) showed linear enhancement of bilateral or unilateral epidural space on axial reformatted images. This finding corresponds to segmental posterior epidural venous dilatation on spinal phlebography (Fig. 1). Three other patients showed prominent epidural enhancement surrounding the thecal sac (cases 1-3). Case 1 appeared to show dural canal narrowing due to the dilated epidural veins (Fig. 2), and case 3 showed unusual anastomosis between the epidural veins and the posterior muscular veins (Fig. 3).

Therefore, cervical epidural venous configurations were roughly classified into two categories; dilatation of posterior epidural veins, and dilatation of all epidural veins.

It was not clear whether the cord atrophy preceded epidural venous dilatation or vice versa, since all patients showed both conditions simultaneously.

At the upper thoracic level, five patients showed prominent posterior epidural veins (cases 3-5, 8, and 10). However, as the posterior epidural space was generally widened at this level, it was difficult to assess whether or not the veins were abnormally dilated.

There was no significant discrepancies between the findings of volumetric MR images and those of conventional phlebography in the 4 cases in which both studies were performed.

The cervical posterior epidural veins and the asymmetry of the spinal cord were more prominent in flexed neck position than in neutral or extended neck position (Fig. 4).

The asymmetry of the internal jugular vein did not bear any correlation with the patients' involved side.

Discussion

In the current study, epidural venous dilatation was observed in all of the patients diagnosed with juvenile amyotrophy of distal upper extremity, who had already shown distal muscular atrophy and focal spinal cord atrophy. These epidural veins are unusual in our limited experience. Hence, this observation strongly suggests that the abnormal epidural vessels are closely correlated with spinal cord atrophy. Moreover, the cases with complete epidural venous dilatation suggests that the problem is not confined to posterior veins.

Several controversial hypotheses have been proposed for the etiology of the epidural venous dilatation: 1) the focal decompression induced by spinal cord atrophy, that is accentuated by neck movement, provokes secondary posterior epidural venous dilatation; 2) the recurrent pressure on the dural canal due to the movement of the bony spinal canal with neck flexion induces venous dilatation. Consequently, the compression induced by dilated veins or congestive ischemic insult incites spinal cord atrophy (12,13); and 3) the dilated vessels present venous malformation (14). Whether the venous dilatation is primary or secondary is a point of concern, although it is difficult to assess even with a histopathological study.

Continuous observation and further discussion of meningorachidian venous plexus in this disorder is necessary. The finding of epidural venous dilatation without spinal cord atrophy, which is considered to be pathognomotic, may be obtained in the early phase of this disease. Since the Gd-DTPA enhanced MR imaging is easier and less invasive than conventional phlebography, it most likely becomes a commonly used option (18).

We should consider three factors which influence the configuration of epidural veins.

First, the flow volume varies depending upon the patient's position, i.e. lying, standing, neck extended or flexed. In standing position, the venous flow from the cranium proceeds dominantly into the epidural veins (19). In neck overflexed position, the flow proceeds into the posterior epidural veins, as the anterior veins are stretched and compressed by the vertebral body. As there are no check valves in the epidural veins (16), the main flow in the anterior longitudinal veins can easily shift to the posterior veins.

Second, in a high flow state, such as epidural vascular malformation or internal jugular vein obstruction, venous drainage exceeds the capacity of the anterior veins and consequently proceeds into the whole epidural system surrounding the dural canal.

Third, the epidural venous flow may change periodically during the disease process. Case 1 showed apparent change of configuration of epidural veins in two MR studies performed in three-month intervals with identical imaging parameters. This case suggests that some other cause besides the patients' position affected the epidural venous flow.

Changes of intrathoracic pressure during the respiratory cycle (17,19) and abnormal posterior lordosis of the spine also affect the epidural veins.

The spinal cord atrophy should be evaluated by MR imaging for the management of a patient with juvenile amyotrophy of distal upper extremity (11). MR studies are also useful for excluding diseases that may possibly be present with similar symptoms, i.e. syringomyelia, arachnoid cyst, epidural tumors and chronic idiopathic demyelinating polyradiculoneuropathy with conduction block (20-22).

The authors believe that the evaluation of epidural veins utilizing enhanced volumetric MR study can be an effective aid in understanding juvenile amyotrophy of distal upper extremity.

Acknowledgement:
We would like to express our thanks to Dr. Heihachiro Aii, Department of Neurology, Otsu City Hospital, for his invaluable advice. We would also like to thank Dr. Hidehiko Nabatame, Department of Neurology, Shiga Medical Center for Adult Disease, for referring one of his patients to us.

Summary
The authors observed epidural venous system in eleven cases with Juvenile
Amyotrophy of Distal Upper Extremity, using spinal phlebography and Gadolinium-DTPA enhanced volumetric MR study. Unusual findings of epidural veins were detected in every case. Further discussion of the findings of epidural veins in this disease is necessary. However, Gadolinium-DTPA enhanced volumetric MR imaging is effective aid for evaluation of epidural veins.

References

1. Hirayama K. Toyokura Y, Tsubaki T. Juvenile muscular atrophy of unilateral upper extremity: a new clinical entity. (in Japanese) Jpn J Psychiatry Neurol 61: 2190-7; 1959.

2. Hirayama K. Juvenile non-progressive muscular atrophy localized in the hand and forearm. -Observation in 38 cases- . (in Japanese) Clin Neurol 12: 313-24 ;1972.

3. Hashimoto O, Asada M, Ohta M, Kuroiwa Y. Clinical observations of juvenile nonprogressive muscular atrophy localized in hand and forearm. J Neurol 211: 105-10; 1976.

4. Sobue I, Saito N Iida M, Ando K. Juvenile type of distal and segmental muscular atrophy of upper extremities. Ann Neurol 3: 429-32; 1978.

5. O'sullivan DJ, Mcleod JG. Distal chronic muscular atrophy involving the hands. J Neurol Neurosurg Psychiat 41: 653-658; 1978.

6. Singh N, Sachdev KK, Susheela AK. Juvenile muscular atrophy localized to arms. Arch Neurol 37: 297-9; 1980.

7. Gourie-Devi M, Suresh TG, Shankar SK et al. Monomeric amyotrophy. Arch Neurol 41: 388-94; 1984.

8. Peiris JB, Seneviratne KN, Wickremasinghn HR, et al. Non familial juvenile distal spinal muscular atrophy of upper extremity. J Neurol Neurosurg Psychiat 52: 314-9; 1989.

9. Oreyma J, Ashby P, Spiegel S. Monomeric amyotrophy. Can Neurol Sci 17: 124-30; 1990.

10. Hirayama K, Tomonaga M, Kitano K, Yamada T, Kojima S, Arai K. Focal cervical poliopathy causing juvenile muscular atrophy of distal upper extremity: a pathological study. J Neurol Neurosurg Psychiat 50: 285-90; 1987.

11. Biondi A, Dormont D, Weitzner I, Bouche P, Chaine P, and Bories J. MR imaging of the cervical cord in juvenile amyotrophy of distal upper extremity AJNR 10: 263-68; 1989.

12. Iwasaki Y, Tashiro K, Kikuchi S, et al. Cervical flexion myelopathy; A "tight dural canal mechanism" J Neurosurg 66: 935-7; 1987.

13. Tokumaru Y, Hirayama K. Anterior shift of posterior cervical dura mater in patients with juvenile muscular atrophy of unilateral upper extremity. Clin Neurol (in Japanese) 29: 1237-43; 1989.

14. Aii H, Koyama M, Nishiura I, Togashi K, Neuroradiological findings

in Hirayama's disease. (in Japanese) Jpn J Clin Radiol 37: 563-8; 1992.
15. Okumura R, Asato R, Miki Y, and Konishi J. Three-dimensional MR Vascular Imaging of the Spine Using Gadolinium-DTPA. Rad Med 10 215-22; 1992.
16. Théron J, Moret J. Spinal Phlebography, Springer-Verlag Berlin Heidelberg New York 1978.
17. Dilenge D, Perey B. An angiographic study of the meningorachidian venous system. Radiology 108: 333-7; 1973.
18. Gelber ND, Ragland RL, and Knorr JR Gd-DTPA enhanced MRI of cervical anterior epidural venous plexus. J Comput Assist Tomogr 16: 760- 63; 1992.
19. Eckenhoff JE, The physiologic significance of the vertebral venous plexus. Surg Gynecol Obstetric 72-8; 1970.
20. Mukai E, Sobue I, Takahashi A, Murakami K and Goto S. Demonstration of central cavity in juvenile-type distal and segmental muscular atrophy of upper extremities. (in Japanese) Clin. Neurol 24: 562-6; 1984.
21. Oka N, Akiguchi I, Kameyama M, Tsuji N, Koyama T. Two cases of cervical intradural arachnoid cyst simulating juvenile muscular atrophy of unilateral upper extremity (Hirayama disease) (in Japanese) Clin Neurol 25: 614-9; 1985.
22. Kitagawa M, Tashiro K, Kikuchi S, and Matsuura T. Diagnosis and differential diagnosis of juvenile muscular atrophy of unilateral upper extremity (Hirayama disease). (in Japanese) Spine & Spinal cord 5:109-14; 1992.

Captions

FIG. 1 Case 6. A 16 year-old male had been aware of distal right forearm motor weakness for three years. a: Axial reformatted images of volumetric study at C6 level, showing bilateral posterior epidural veins (arrows). b and c: Spinal phlebography performed three weeks after MRI clearly showed the segmental dilatation of bilateral posterior epidural veins [right side (b), left side (c)]. d: Enhanced CT image at C6 level with neck flexion showed enhancement of posterior epidural space, suggesting dilatation of the posterior epidural veins. The difference in posterior epidural space between a and d was most likely a result of the patient's posture. This MR finding may be difficult to identify without having first analyzed the referenced image of spinal phlebography. Gd-enhanced MR volumetric study can clearly show the posterior epidural veins without requiring the patient to be in neck flexed position.

FIG. 2. Case 1. An 18 year-old male had insidiously noticed right hand weakness a year and two months before consulting a neurologist because of dorsal right hand amyotrophy noted four months prior to the consultation. a and b: Axial reformatted images of volumetric study (performed 2/26/92) showed unusually high epidural venous flow in the anterior longitudinal veins at the C2/3 level (a), dispersing into vessels surrounding the thecal sac, most notably the posterior veins at the C6 level (b). Narrowing of the spinal dural canal was also apparent. c: Spinal phlebography performed one week later verified the MR findings. d: A sagittal enhanced T1-weighted image with fat suppression (performed on 2/26/92). e: A sagittal T1-weighted image (performed on 12/5/91). Apparent discrepancy of epidural vessel configuration was noted on both images, although the patient's posture was almost identical. This case strongly suggests that the condition of the epidural veins may change periodically.

FIG. 3. Case 3. An 18 year-old male had slowly developed right hand weakness over a 1 year period. a: Axial reformatted images showed unusual anastomosis between the epidural vein and the dorsal muscular vein at the C3/4 level (white arrows). b: Sagittal projection images. The left and right images were obtained by collapsing three

selected contiguous parasagittal images of the patient's left and right sides, respectively. Although the clinical significance of this finding is unclear, it is a unique radiological finding which can only be obtained by the present method.

FIG. 4. Case 10. A 19 year-old male had slowly developed right hand weakness over a 4-year period. a: a sagittal Gd-DTPA enhanced T1 weighted image and b: an axial T1-weighted image at C6 level in neutral neck position. c: a sagittal post contrast enhanced T1-weighted image and d: an axial image at C6 level in neck flexed position. Posterior epidural space as well as the enhancement of veins is more prominent in flexed neck position, probably due to the flow shift from the anterior epidural system to the posterior system. Although the hemiatrophy of the spinal cord is apparent in flexed neck position, the authors believe that this finding does not necessarily suggest spinal cord congestion.

Fig. 1 (a)

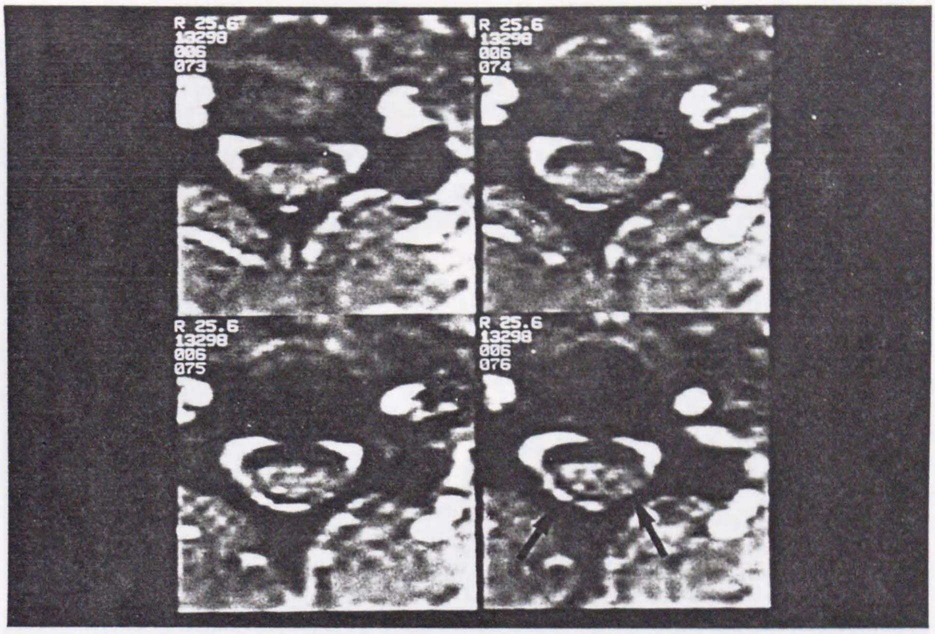


Fig. 1 (b)

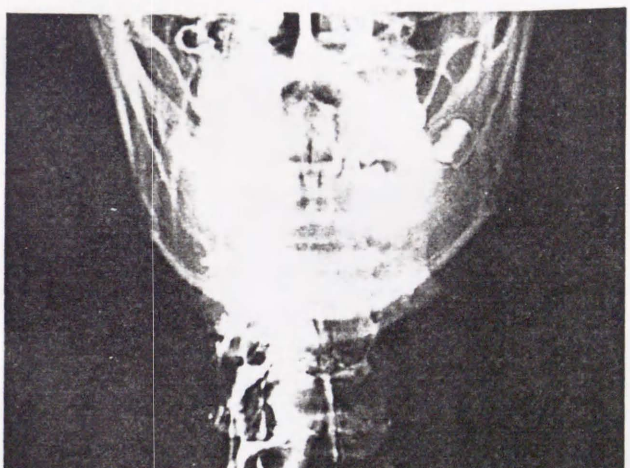


Fig. 1 (c)

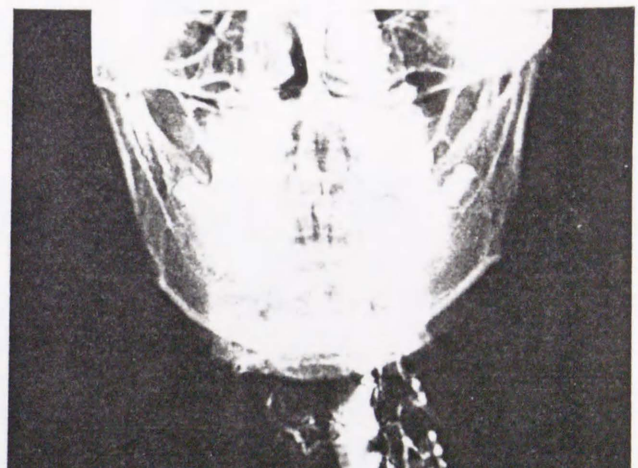


Fig. 1 (d)

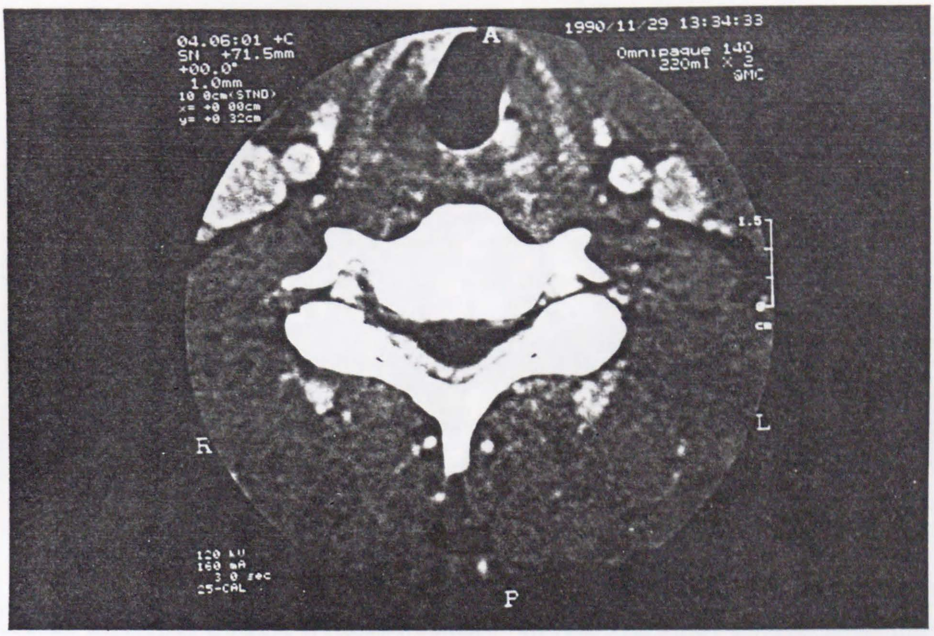
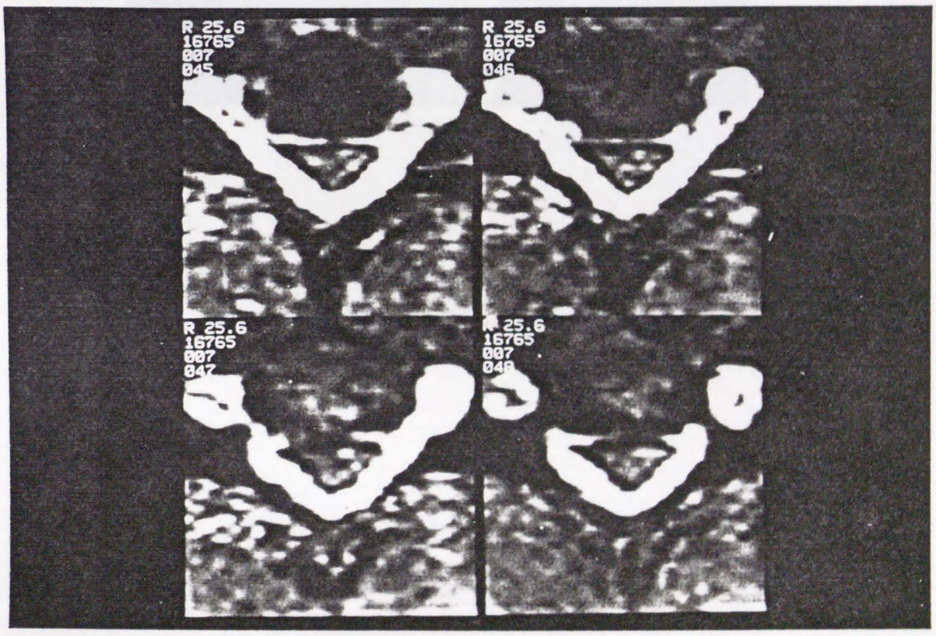

04.06:01 +C
SN +71.5mm
+00.0°
1.0mm
1990/11/29 13:34:33
Omnipaque 140
220ml X 2
A
R
L
P
1.5
cm
120 kV
160 mA
25-CAL

Fig. 2 (a)

007
017
16765
007
018
16765
007
019
R 25.6
16765
007
020

Fig. 2 (b)



R 25.6
16765
007
045
R 25.6
16765
007
046
R 25.6
16765
007
047
25.6
16765
007

Fig. 2 (c)

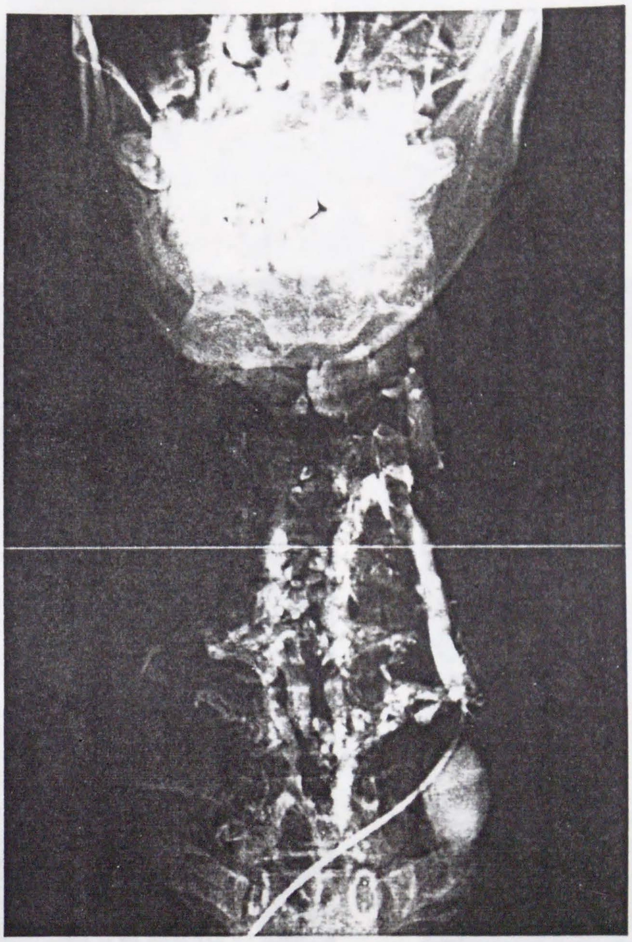


Fig. 2 (d)


: 16765
: 005/06
: 005/009
S120
R 1
I120

Fig. 2 (e)


MAGNETOM 1 5 T
SHIGA SEIJINBYO CE
18 M
DI E
UP
MR-4911
05-DEC-91
13 52:00
STORE>12
TR 55
TE 15
SL 4 0
SP 1 6
X 0
ZF 1 20
W
C
TA

Fig. 3 (a)

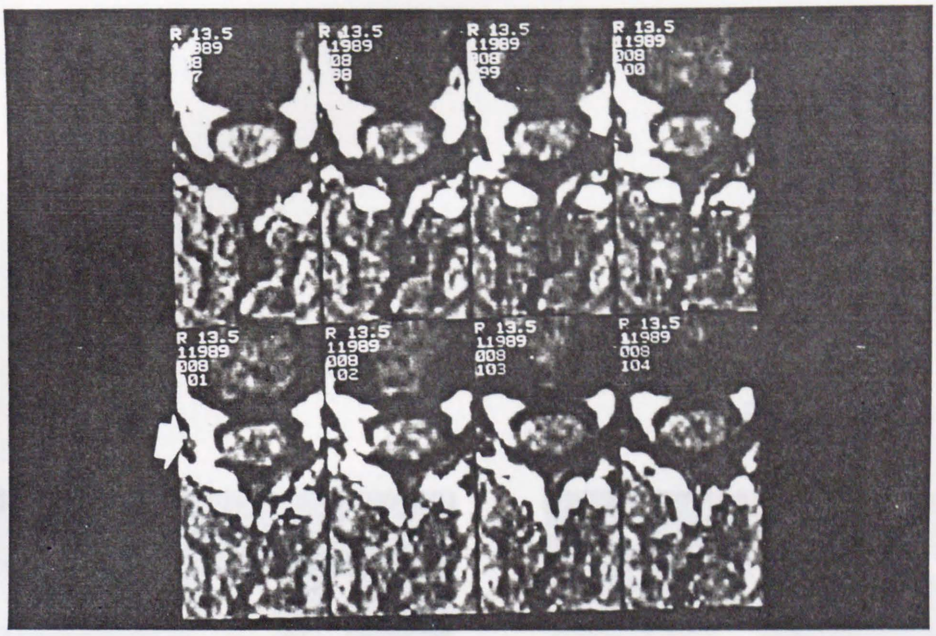


Fig. 3 (b)

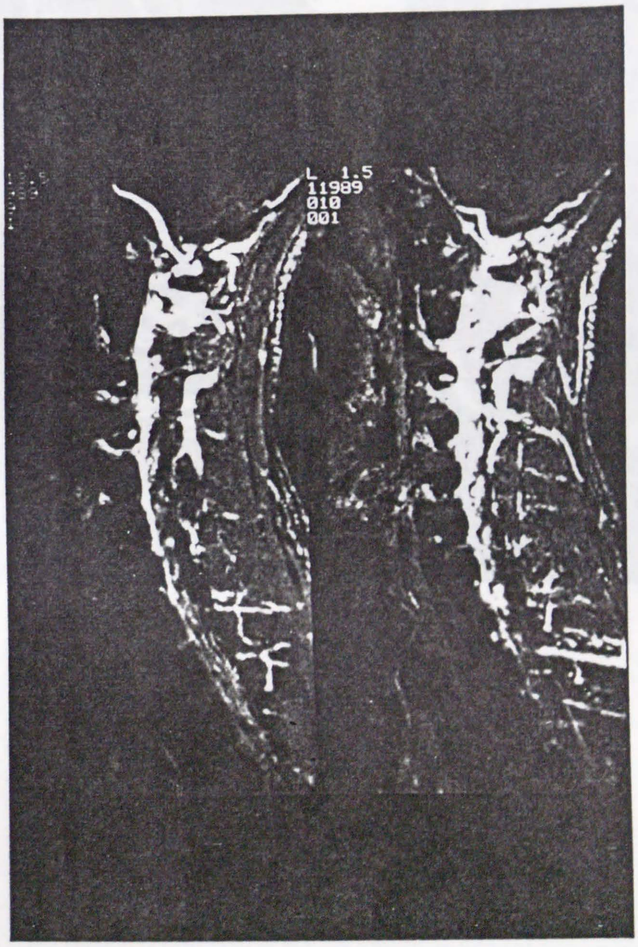


Fig. 4 (a)

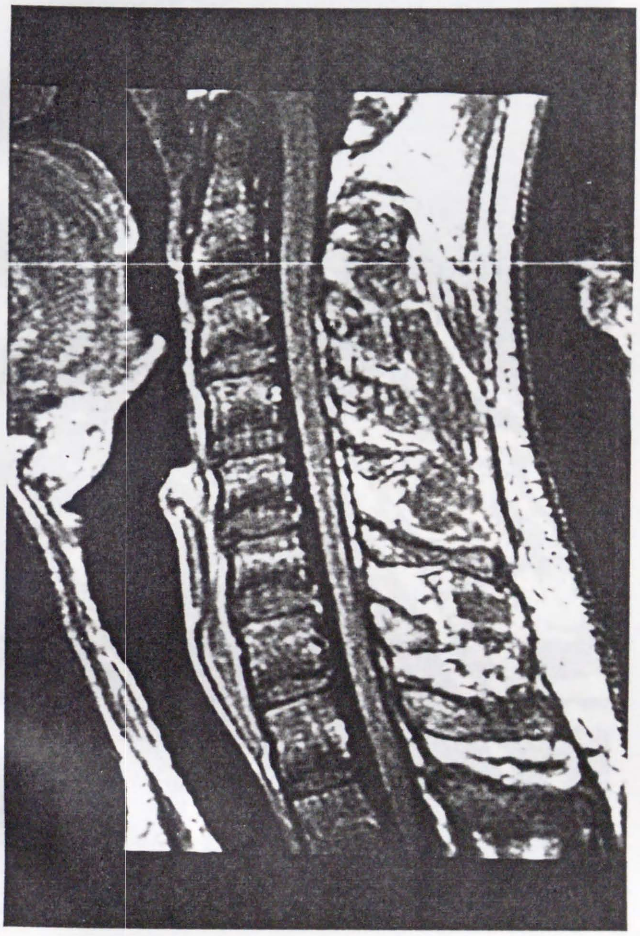


Fig. 4 (b)

Fig. 4 (c)

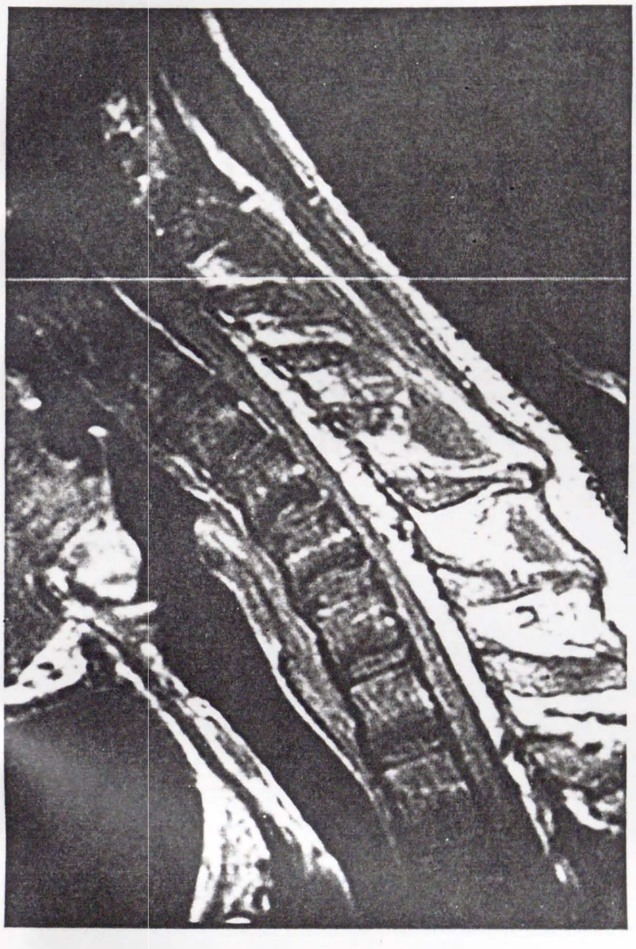


Fig. 4 (d)

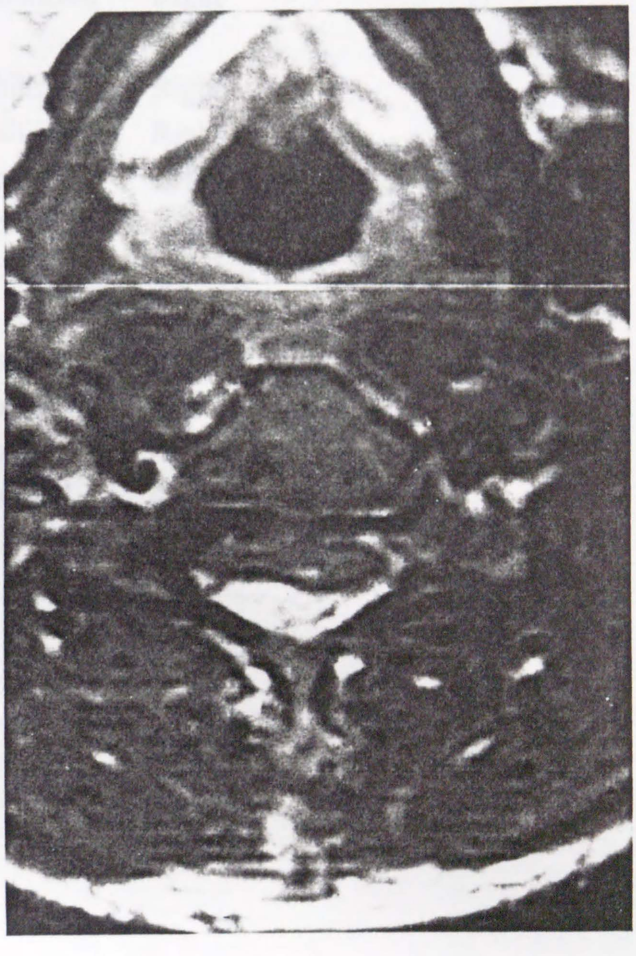


TABLE 1. Patients' data

Case No.	Age	Sex	Duration of disease	Disease phase	Side Involved
Case 1	18	M	1y4m	Stabilized	Right
Case 2	22	M	6m	Evolutional	Right
Case 3	18	M	3y	Stabilized	Right
Case 4	17	M	1y	Stabilized	Right
Case 5	26	F	2y	Stabilized	Left
Case 6	16	M	3y	Stabilized	Right
Case 7	19	M	3y	Stabilized	Left
Case 8	48	M	33y	Stabilized	Right
Case 9	19	F	4y	Stabilized	Left
Case 10	19	M	4y	Stabilized	Right
Case 11	28	F	2y	Stabilized	Right

TABLE 2. MRI and phlebographic findings

Case No. /Involved side	Spinal cord atrophy	Int. jugular veins	Spinal phlebography	Epidural venous configuration shown on volumetric MRI
Case 1 /Right	Right hemiatrophy	Left small	Prominently dilated	Prominent epidural veins surrounding the thecal sac (cervico-thoracic level)
Case 2 /Right	Right-dominant bilateral atrophy		Left slow flow	Dilated Prominent epidural veins surrounding the thecal sac (cervico-thoracic level)
Case 3 /Right	Right hemiatrophy	Symmetrical	Uncatheterized	Anastomosis between the right dorsal epidural and muscular veins at the C3/4 level, Prominent anterior epidural veins (Right dominant, cervical level), Posterior epidural veins (thoracic level)
Case 4 /Right	Right hemiatrophy	Right small	Not performed	Curvilinear posterior epidural veins (C 5-6 level) Posterior epidural veins (thoracic level)
Case 5 /Left	Left hemiatrophy	Symmetrical	Mild dilatation of posterior epidural veins	Curvilinear posterior epidural veins (C 5-6 level) posterior epidural veins (thoracic)
Case 6 /Right	Bilateral atrophy	Symmetrical	Segmental dilatation of posterior epidural veins	Curvilinear posterior epidural veins (C 5-6 level)
Case 7 /Left	Left hemiatrophy	Left small	Not performed	Curvilinear posterior epidural veins (C 5-6 level))
Case 8 /Right	Bilateral atrophy	Symmetrical	Not performed	Curvilinear posterior epidural veins (cervical level) Posterior epidural veins (thoracic level)
Case 9 /Left	Left hemiatrophy	Symmetrical	Not performed	Curvilinear posterior epidural veins (C 5-6 level))
Case 10 /Right	Right hemiatrophy	Symmetrical	Not performed	Curvilinear posterior epidural veins (cervical level) Posterior epidural veins (thoracic level)
Case 11 /Right	Right hemiatrophy	Symmetrical	Not performed	Right curvilinear posterior epidural veins (cervical level)